

Delayed Diagnosis of Congenital Hearing Loss in Preschool Children

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THE CARDINAL HANDICAP of the infant or preschool child with congenital hearing loss is delay in the development of language. The resultant poor comprehension and expression of language blight the growth of the young mind, with secondary restriction of mental and social expression. If habilitation is to be effective, it must begin during the child's first and second years, when language is normally learned, and must seek to bring residual hearing into use before the infant develops an auxiliary nonverbal communication system; gesturing, for example. The early diagnosis of hearing loss thus is essential for the development of a program of habilitation, and the important role of the medical profession in this area of child health is readily evident.

All too often the recognition and appropriate assessment of infants and children with congenital hearing loss are needlessly delayed. Efforts to reduce such a time lag are extremely important in the education of these children, indeed in the foundation of their lives. This

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study was undertaken to review the pattern of referral of preschool children with congenital hearing loss enrolled in the provincial preschool hearing program in British Columbia (1) and to determine the effectiveness of public health contacts as an aid to the early recognition of this handicap. The results of the study provided clues for the improvement of casefinding by the public health service.

British Columbia, the westernmost Province of Canada, has an area of 366,255 square miles and a population of approximately 1.75 million. Its public health services are provided by a number of health units comprising two metropolitan areas, Vancouver and Victoria, and 17 provincial units. Each health unit has its own director and regional program, but all units are coordinated by the provincial department of health.

The metropolitan and provincial health unit boundaries are shown in figure 1, together with the number of children enrolled in the provincial preschool hearing program from each health unit in British Columbia. More than half of the population is concentrated in the southwest corner of the Province, and a little more than half of the caseload is in this area.

Methods and Materials

The study material was derived from the first 300 children seen in the program. The children were examined in the outpatient department of the Health Centre for Children at the Vancouver General Hospital. No child with an average hearing level of less than 30 decibels in the

better ear was included in the series. More than 65 percent of the children had hearing loss in excess of 60 decibels above speech pressure level (1).

Thirty-four children whose hearing loss was caused by a postnatal illness were excluded from the study. The names of the remaining 266 were submitted to the metropolitan health departments in Vancouver and Victoria and to the British Columbia Provincial Health Branch. In turn, they distributed the names to the regional health units throughout their areas of administration. An additional 30 children were excluded either because they could not be located during the field study or because the report concerning them arrived too late to be included in the analysis. Satisfactory information was available on 236 of the 300 children.

The health unit staffs were asked to review their records to see whether there had been a public health contact with the family and particularly whether a child health conference had been visited. In the 130 cases with a contact, the public health nurses were asked whether the hearing loss had been suspected.

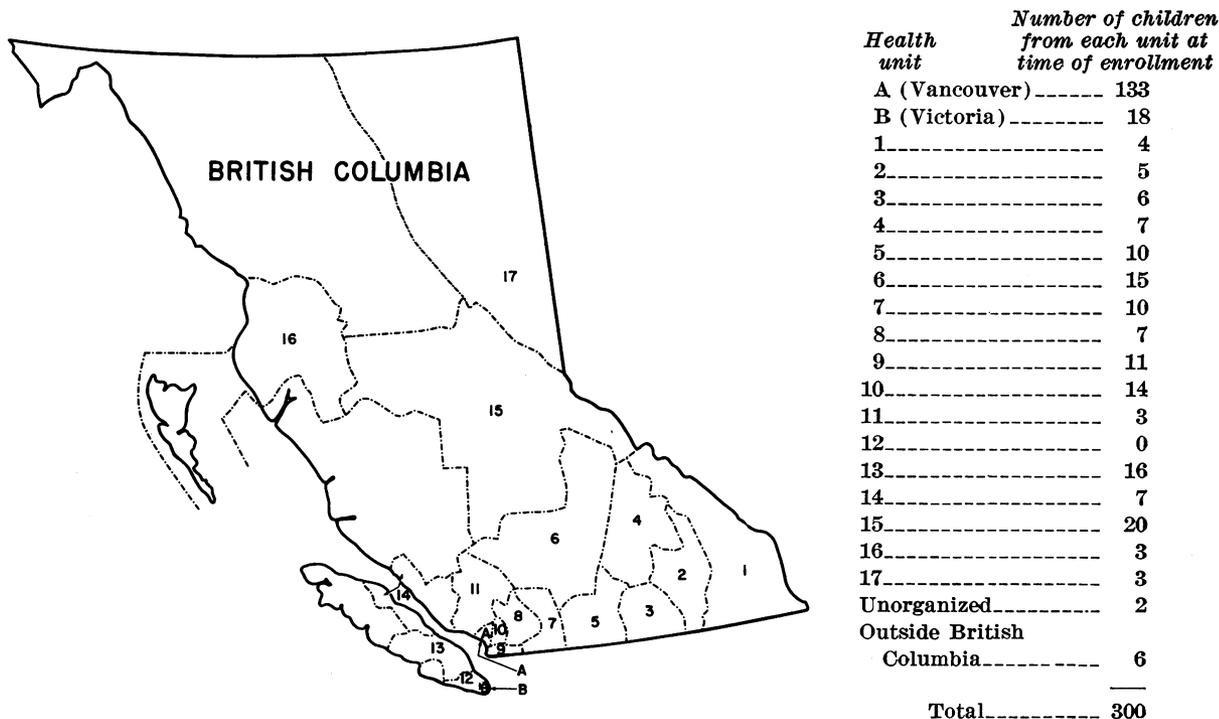
Results

Age hearing loss suspected and enrollment. The age of the children when hearing loss was suspected by the families and the age at enrollment in the preschool hearing program at the Health Centre for Children are compared in figure 2. Only 33 percent of the cases were suspected by the families during the first year of life; 89 percent of the cases were suspected during the first 3 years of life. The delay in the referral of these children is evident.

The child's age when enrolled at the Health Centre for Children shows that the pattern of late referral was similar for both the metropolitan and provincial areas (fig. 3). Only 23 percent of the 236 children were referred during the first 2 years and 52 percent during the first 3 years of life. The maximum number were enrolled during the third year of life.

The severity of hearing loss for each age group is shown in figure 4 for the combined metropolitan and provincial groups. There was a trend for profound and severe cases to be referred earlier than moderate and mild cases, but a distressing number of children with im-

Figure 1. Health unit boundaries in British Columbia, 1960, and number of preschool children enrolled in hearing program in each health unit



portant degrees of congenital hearing loss were not enrolled until 4 to 6 years of age.

Public health contact. In the two metropolitan areas, Vancouver and Victoria, less than half of the families and their children had a public health contact, whereas 75 percent had a contact in the provincial areas (table 1). All contacts in Vancouver, 2 of 9 in Victoria, and 43 of 80 in the provincial health units were in child health conferences.

The number of children suspected of hearing loss at the public health contact are given in table 2. Records show that 39 percent were suspected of hearing loss in Vancouver, none in Victoria, and 31.2 percent in the provincial health units. Of the 130 children contacted, only 41 (31.5 percent) were suspected of hearing loss at contact.

Family mobility. The mobility of family groups has an important influence on the continuity of long-term care of children with severe handicaps of this type. The number of moves for this group of children are given in table 3. Approximately one family in five moved from

Figure 2. Age when parent suspected hearing loss and age at enrollment in hearing program of 236 preschool children, British Columbia

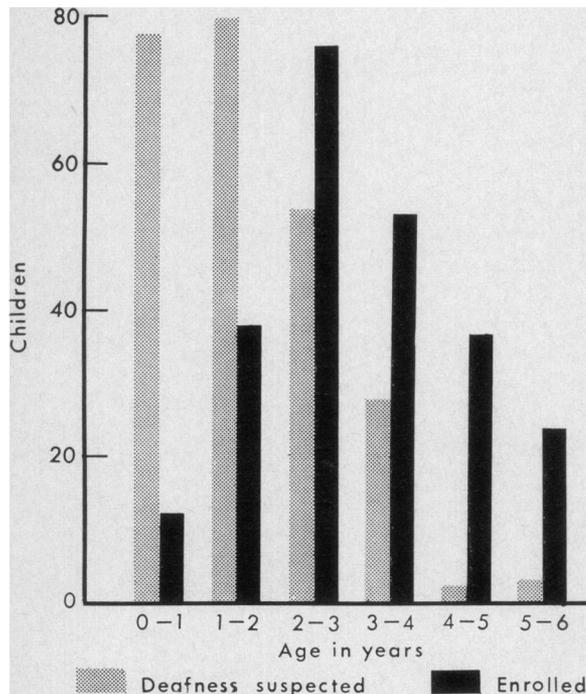
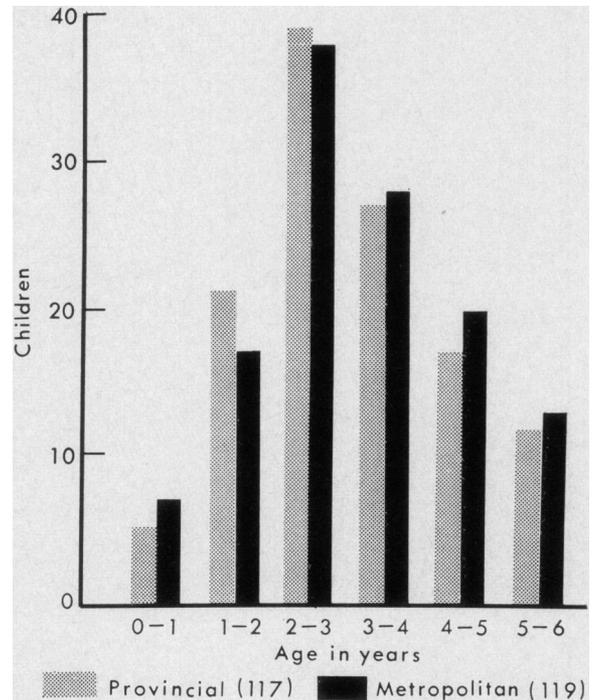


Figure 3. Age at enrollment in hearing program of 236 preschool children from provincial and metropolitan areas, British Columbia



one health unit to another within the Province after enrollment at the Health Centre for Children.

Mortality. Three children in our study series died from accidental causes: two from traffic accidents and one from burns in a house fire.

Comment

Our data on age of child when the family suspected a hearing loss and on age at enrollment in the preschool hearing program are comparable to those reported by the University of Kansas Medical Center (2). They noted parental suspicion of hearing loss in 40 percent of the children (16 of 39) by the end of the first year and similar delays in referral for assessment, with the peak during the third year of life (26 of 42). The recent report of the Carnegie United Kingdom Trust (3) on the problems of 600 handicapped children and their families noted the delay in suspicion of deafness which was aggravated by delay in referral for specialist investigation. Parental suspicion of hearing loss cannot be relied upon to

insure early detection in infants and young children.

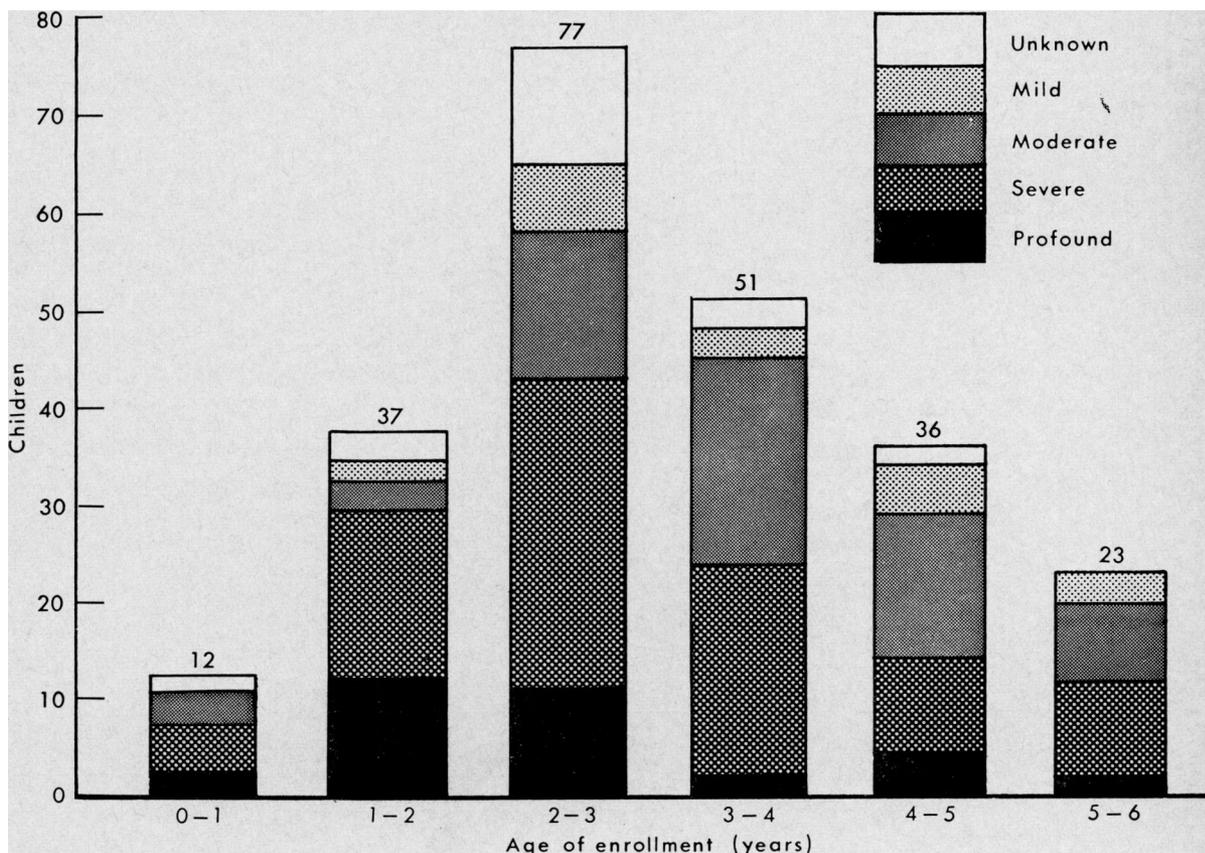
Public health contacts in British Columbia also have been ineffective in finding children with an important degree of hearing loss early in their lives. When it is considered that nearly half of the children from the metropolitan areas and 75 percent of the children from the provincial areas had a public health contact, it is clear that the casefinding potential of this facility has been overlooked. The delayed enrollment in the hearing program until from 4 to 6 years of age of the children in our study series with severe and profound hearing loss underlines a prevailing ignorance of the urgency for early detection and early training.

As a result of our study, an inservice training session was organized for public health medical and nursing personnel by the medical and audiological staff of the hearing and language clinic in the Health Centre for Children.

At this session the physician noted that parents suspected hearing loss for two reasons, (a) lack of response to sound and (b) delay in speech development, and he stressed the importance of a high level of suspicion of hearing loss. He further explained that understanding the known causes of hearing loss increased the likelihood of early detection, and he considered all infants and children at risk when there was a history of genetic deafness, maternal rubella during the first trimester, neonatal anoxia or hemolytic disease of the newborn, meningitis, or exposure to ototoxic drugs. In our experience the cause of hearing loss has been known in 50 percent of the cases (4).

The audiologist demonstrated techniques of evaluating hearing in infants and small children. The need for a quiet room for testing—obvious although not always available—was mentioned. Various uncalibrated noisemakers were recommended for use by the public health

Figure 4. Age at enrollment in hearing program of 236 preschool children, by severity of hearing loss, British Columbia



nurse. These included a high-pitched rattle, a bell, squeaky toys, a clicker, the crunch of tissue paper, the movement of a spoon on the rim of a cup, and loud and soft voices, with breath and vibration screened out with paper. Testing is carried out with the baby, on the mother's lap, facing an assistant who attracts the baby's attention and observes facial expression and movements. The tester and testing equipment are hidden from the infant's vision and the sounds made well to each side of the baby and not above or below the head. Nurses were cautioned to avoid air vibrations or perfumes, which could provide clues for a baby with hearing loss. If the baby failed to react to sound, the nurse was advised to wait 2 seconds and to repeat the sounds.

With a case history to search for high-risk babies, this screening technique should aid in the selection of a group requiring referral for further hearing evaluation. It is still too early to know the results of this session, but a greater awareness on the part of the public health staff has already resulted in the early identification of several children with varying degrees of hearing loss.

This study of children with hearing loss exposes a major problem in contemporary child health in our society—the delayed recognition of the handicapped infant that prejudices and even nullifies the habilitation effort. Current developments in the early diagnosis of neuro-metabolic disease (5) offer yet another example of the urgency of identification and result from a somewhat belated acknowledgement of the prevalence of these serious handicaps and their ultimate cost to society.

The importance of early identification of seri-

Table 2. Number of children suspected of hearing loss at public health contact among 236 preschool children, British Columbia

Area	Children with hearing loss	Number with contact	Suspected at contact	
			Number	Percent
Vancouver.....	111	41	16	39.0
Victoria.....	19	9	0	-----
Provincial.....	106	80	25	31.2
Total.....	236	130	41	31.5

Table 3. Number of children with a public health contact who have moved from one unit to another within British Columbia

Area	Children with hearing loss	Children who moved	
		Number	Percent
Vancouver.....	111	20	18.0
Victoria.....	19	2	10.5
Provincial.....	106	22	20.8
Total.....	236	44	18.6

ous handicaps in children is indicated in the study by Bierman and associates (6). They studied the incidence of physical and mental handicaps of prenatal and natal origin and classified them into four classes according to their impact on the community and the type and duration of care required. They estimated that of the 10 percent of liveborn infants requiring special care, almost two-thirds were amenable

Table 1. Number of children with a public health contact among 236 preschool children with congenital hearing loss, British Columbia

Area	Children with hearing loss	With contact		Without contact		Unknown	
		Number	Percent	Number	Percent	Number	Percent
Vancouver.....	111	41	36.9	56	50.5	14	12.6
Victoria.....	19	9	47.4	10	52.6	0	-----
Provincial.....	106	80	75.5	23	21.7	3	2.8
Total.....	236	130	55.1	89	37.7	17	7.2

to short-term skilled medical and nursing care. The remainder, almost 4 percent of all the live-born infants, required combinations of diagnostic services, long-term skilled treatment, special therapy, special education, and custodial care; and even with the best of care, three-fourths of these children would be permanently handicapped to some degree.

This small group of children constitutes a major social burden to the community, and infants and young children with hearing loss, with or without other abnormalities, represent an important segment of this group. The control and partial cure of previously fatal diseases of the neonate and infant (prematurity, hemolytic disease of the newborn, neonatal anoxia, and meningitis) have contributed to the increasing pediatric population with multiple handicaps, a population whose continuing care requires early casefinding, comprehensive assessment, and planned programs of rehabilitation.

Advances in medical care have created many new problems—the paradox of medical progress (7). One is the existing need for an organized search for high-risk infants, in nurseries and throughout the first years of life, and their prompt referral to good diagnostic facilities. Risk registries (8) are an interesting development to effect the early recognition and assessment of handicaps and stress the idea that the search be concentrated on a population of high-risk children. Estimates of children at risk vary from 10 to 20 percent of live births. The most profitable use of such risk registries has been found in the detection of every type of handicap rather than any single handicap, such as deafness. Sheridan (8) considers that these children fall into five main categories: those with an unfavorable family history; the three groups who have been exposed to adverse environment in prenatal, perinatal, or postnatal life; and a symptomatic group whose development is in any way retarded or abnormal. Rossi (9) has defined a high-risk baby as one who stands a greater than average chance of developing a neonatal life-threatening disease or sequelae, due usually to maternal disease or complications at birth.

It has been suggested that the most suitable person to compile and maintain such a register

is the medical officer of health. The family physician cannot cope singlehandedly with the planning necessary to insure early identification of all high-risk infants, followed by assessment and arrangements for continuing followup, because the mobility of families—and even physicians themselves—undermines the continuity of care. A clear-cut need exists for regional health departments and physicians interested in child health to cooperate in developing case-finding programs and perhaps risk registries. It is also reasonable that the administrative machinery and knowledge of community facilities of the health department be available to the family physician to expedite referral and followup of selected children with serious handicaps.

The successful development of risk registries on a local basis depends on the close cooperation of the family physician and the public health department. Their use may permit identification of hearing loss when the physician makes his postnatal followup visit, when the public health nurse visits the home as early as 4 to 6 weeks after delivery; and certainly at 3 to 6 months in the child health conference. In British Columbia, a copy of the "Notification of Birth" form is made available to each health unit, where the relevant information concerning the prenatal and early perinatal history of mother and infant is theoretically available for scrutinizing by public health personnel before contacting new babies.

The British Columbia registry for handicapped children and adults (10) was organized in 1948 to obtain accurate morbidity statistics of crippling diseases in children and, subsequently, adults; to assist in followup of individual cases; and to demonstrate the need for added services for the handicapped. Such a central registry is not designed to identify the handicapped (11).

There is widespread agreement that children with congenital hearing loss should be enrolled in a special educational program during the years of life when language is normally learned. The teacher of the deaf can do little to expedite the early referral of pupils. This is the job of the medical and allied health professions. Family physicians and public health personnel

must be alerted repeatedly to the need for a routine and deliberate search for these infants and young children.

Summary

A study of the effectiveness of a public health contact in casefinding was undertaken in a series of 236 children enrolled in a preschool hearing program in the Province of British Columbia. A total of 130 children had a public health contact, either when the home was visited or at a child health conference. Hearing loss was suspected in only 31.5 percent of the group at this contact.

When it is considered that nearly half of the children from the metropolitan area and three-fourths from the provincial area had a public health contact, the casefinding potential of the public health service can be considered impressive.

Some methods are outlined to improve early casefinding by public health personnel and family physicians. Maximum effectiveness of such a program depends on close liaison between family physician, public health agency, and special diagnostic facilities.

Early medical recognition is needed to insure optimal habilitation of children with congenital hearing loss of sufficient degree to interfere with the development of language. It is well known that an unnecessary delay often occurs in referring these children for proper assessment and the initiation of habilitation.

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